CASE AND COMMENT

FUNDOSCOPY

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CASE

A general medical officer assigned to a federal health program in a relatively remote area evaluated and treated, medically, an 11-year-old girl with a history of episodic headache that was considered consistent with migraine. The family history, in multiple maternal relatives, was remarkably positive for migraine.

Initially, the child appeared to respond quite well. Subsequently, however, the patient's mother notified the physician that the girl's headache had become persistently intense, without response to medication. Further, the mother noted that the girl had become anorexic, with weight loss, and that she had been vomiting. Admission to a local dispensary-hospital was arranged. Nurses in attendance documented that the child upon presentation was confused, listless, and ataxic. Following his evaluation of the patient, the attending physician determined that transfer was necessary to a nearby medical center, where specialty consultants and specialized diagnostic facilities were available. Given the limitations of pertinent medical records, fundoscopy at that time was either not performed or considered normal.

Because the physician considered an infectious process as his primary working diagnosis, lumbar puncture for analysis and culture of cerebral spinal fluid was performed prior to the institution of intravenous antibiotics. During transfer, the patient experienced respiratory arrest. Upon arrival at the receiving hospital, findings included fixed dilated pupils with fully developed bilateral papilledema and retinal hemorrhages. A CT scan shortly thereafter revealed a large parietal lobe tumor and evidence of bilateral subtentorial herniation. An emergency craniotomy was performed.

In time, a malpractice claim was filed with the federal government regarding the medical care rendered prior to transfer.

COMMENT

As is true for physicians practicing in a number of specialties, the ability to detect a swollen optic nerve head by fundoscopy is a clinical skill that must be part of the diagnostic armamentarium of any general medical officer. The potential for realizing truly dire adverse clinical outcomes, in the absence of the ability to skillfully evaluate the ocular fundi, should be obvious.

Today, it is the custom of neurologists and neuro-ophthalmologists to reserve the term papilledema solely for those cases of optic nerve head swelling (Table 1) that occur in association with and caused by increased intracranial pressure. Authorities have noted that the differential diagnosis of optic nerve head swelling may be literally endless. In that regard, the differential reproduced as a table to accompany this article derives from several of the references cited and is necessarily an abbreviated summary only.

Differential Diagnosis -
Optic Nerve Head Swelling

1. Papilledema
2. Disc Anomalies
3. Disc Tumor
4. Orbital Tumors
5. Inflammatory Disease
6. Metabolic/Endocrine Disease
7. Intraocular Disease
8. Vascular Disease
9. Systemic Disease

TABLE 1

The recognition of abnormal findings upon ophthalmoscopic examination of the optic nerve head, similar to the development of other basic clinical skills, rests

squarely upon detailed knowledge of, repeated exposure to, and empiric familiarity with normal findings. The attributes of the nerve head to be critically appreciated include its color, the surface topography or contour of the entire nerve head and the physiologic cup, recognition of the margins of the disc, the character of the retinal nerve fiber layer immediately adjacent to each quadrant of the disc, and an assessment of retinal vessels, especially the presence of central venous pulsations along with the clarity of the view of the vessels at the disc margin and within the peripapillary retinal nerve fiber layer.

Although microanalytical debate continues in some quarters regarding the exact pathophysiology of papilledema, a literal misnomer, the final common pathway minimally involves transmission of increased intracranial pressure along the intraorbital optic nerve sheath, an extension of the dura, with a consequential obstruction of normal axoplasmic flow at the lamina cribrosa, the multiply fenestrated window in the sclera behind the nerve head that allows for egress of the axons of the retinal nerve fiber layer from the globe. Intraocularly, the more proximal axons on the surface of the disc, at the disc margin, and within the peripapillary retinal nerve fiber layer become swollen. Glial cells are not involved. Eventually, true interstitial fluid may collect, with subretinal fluid pooling. Another component in the pathophysiology of papilledema is vascular congestion. This may lead, in series, to an obstruction of central retinal venous flow, capillary dilation, breakdown of the normal blood-retinal barrier with exudation or hemorrhage, and the ultimate potential for ischemia with microinfarction.

It is clinically beneficial to divide findings of acute papilledema into an early versus late or fully developed phase. The presence of spontaneous venous pulsations at the disc is consistent with a conclusion that intracranial pressure is normal, i.e., less than 200 - 210 mm of water. Due to the fact that 20% of normal persons have no spontaneous venous pulsations, the absence of such a finding is not a diagnostic criterion.

The findings of papilledema in its most early acute phase (Table 2) can be limited and rather subtle. The temporal optic nerve head, that facing the macula, and the physiologic cup remain entirely normal in appearance. Faint swelling of the axons on the surface of the nerve and/or at its margins may be detected only at either vertical pole. Vascular findings are limited to those of early congestion, with some prominence of the normal capillaries on the disc surface and a telltale change in disc color to a more florid pink or red. It may be, when possible, that diagnosis at this stage can only be attained by repeat observations or upon securing the evaluations of specialty consultants.

Acute Papilledema - Early

1. Normal Temporal Disc
2. Normal Physiologic Cup
3. Vertical Poles - Pinker/Red
4. Vertical Poles - Hazy Margins
5. Vertical Poles - Elevated Axons
6. Vertical Poles - Obscured Vessels

TABLE 2

Fully developed acute papilledema (Table 3) includes prominent elevation of the axonal substance of the disc, along with a noteworthy redness in color, throughout all quadrants of the nerve head. The physiological cup is also elevated and may be compromised, but it is not characteristically abolished. The margins of the disc are faint, and the course of the blood vessels at the margins and within the peripapillary retina are obscured. Hemorrhages may overlie the disc, and the peripapillary retinal nerve fiber layer can be both elevated and thrown into folds. This zone of the retina also includes various superficial

Acute Papilledema - Late/Fully Developed

1. Florid Redness
2. Elevation of Disc Tissue/Axons
3. Encroachment Upon Elevated Physiologic Cup
4. Obscured Margins
5. Obscured Vessels
6. Hemorrhages
7. Nerve Fiber
8. Nerve Fiber Layer Infarcts (Cotton Wool Spots)
9. Nerve Fiber Layer Swelling/Elevation/Folds

TABLE 3

hemorrhages, often by anatomic necessity with a tethered flame appearance, hard exudates, and larger, softer appearing microinfarcts or cotton wool spots.

Chronic papilledema lacks hemorrhages, exudates, and microinfarcts. The color of the disc evolves from florid red to a clouded gray. Tiny dot concretions may be detected within the superficial disc tissue. Fully resolved papilledema may lead to optic atrophy. Axonal loss with capillary dropout and the persistence of glial cells give the nerve head a characteristic bright white-yellow coloration, in either a zonal or complete distribution.

Typically, papilledema arises bilaterally, and its presentation includes the preservation of normal visual acuity. As a working diagnosis, almost by textbook definition, papilledema should be clinically construed as a medical emergency, and noninvasive central nervous system neuroimaging, by CT scan or MRI, should be urgently secured.

A number of the references cited were specifically chosen because they include, often in color, clear reproductions and photographs of the various stages of papilledema, along with similar depictions of examples from the differential diagnosis of optic nerve head swelling.

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